Introduction

GARDNER (2) has shown the association between syringomyelia and abnormalities at the foramen magnum and improvement after craniovertebral decompression, not only for symptoms of the craniovertebral malformation but also those of the associated syringomyelia.

In a series of 118 patients (10) possible causes included spina bifida (6), meningitis (3) and cerebral tumour (2). Other possible factors were severe head trauma in 21 cases and history of difficult birth in 62. Head trauma has been suggested as a causative factor by earlier authors including HERTEL and HILD (3) but the relevance of such an incident is difficult to determine. Head injury at any age may through cerebral anoxia and swelling, sub-arachnoid bleeding, hydrocephalus or basal arachnoiditis cause herniation of the cerebellum. If injury takes place during birth moulding of the skull and overlapping of bones of the vault permit compression of the brain. Invagination or occipital osteodiastasis around the foramen magnum may occur because the expulsive force of the uterus is applied to the base of the skull by the cervical spine. The finding that 62 out of the 118 patients reported some factor predisposing to injury in their birth was of interest. Results of this enquiry and comparison with 200 controls have been published (10).

CHIARI (1) suggested that the hind brain hernia in both the CHIARI type I deformity and the CHIARI type II was due to hydrocephalus, although it is now known that the CHIARI type I malformation may commonly occur without it. In a recent series of 80 patients with syringomyelia in whom ventricular size was assessed by WEST and WILLIAMS (6) 54 were normal and only 3 showed severe hydrocephalus. Symptoms of high pressure in the head are rare in syringomyelia, and it seems that pressure differences across the foramen magnum may be intermittent (7).

In a study of CHIARI type II malformation it has been noted (8) that the important differences in pressure were those in which the intracranial pressure was higher than the intraspinal; this has been called craniospinal dissociation. Pressures in the tissues are important but cerebrospinal fluid (CSF) pressures were more accessible to measurement, therefore only CSF pressure was considered.

Four types of pressure recordings were found:

1. Normal: In these the baseline pressures were equal in the head and the spine (baseline pressures refers to the average pressure ignoring the pulsations due to heart beat, coughing, crying and so on).
2. Baselines equal with valvular effect: In these cases the head pressure rose higher than the spinal pressure but equalised at rest after a few seconds.

3. Baseline dissociation with valvular effect: pressure differences between the head and spine were present at rest and were aggravated by exertion.

4. Total dissociation: In this state, always associated with rapidly progressive hydrocephalus, the pressure events in the head were separate from those in the spine, indicating a watertight blockage at the foramen magnum.

The present results concern a study of adults only.

Materials

Over 100 adult recordings of intracranial and intraspinal pressure measurements gave adequate normal values. Also included were 43 patients with proven syringomyelia in whom 83 recordings have been done of these 38 patients have had preoperative studies and 36 postoperative (31 both pre- and postoperative).

Methods

Ventricular puncture was made through the right frontal twist drill hole (4) using an 8 cm JEFFERSON cannula. Pressure recordings were made from CSF in the sitting position (Fig. 1) and from a mouthpiece.

The pressures were recorded with the patients at rest, during QUECKENSTEDT'S manoeuvre, during coughing and during a modified VALSALVA manoeuvre produced by blowing into the mouthpiece.

Results

In 32 cases the results were normal. In the majority of abnormal cases after CSF had been forced from the spine into the head by the epidural veins, it was delayed by valvular action at the foramen magnum and then slowly returned to the "baseline equal" state. This was best seen after two types of event.

Coughing: After a sharp rise in abdominal pressure such as a cough the pressure rises faster, higher and sooner in the spine than in the head and during recovery the CSF drops back rapidly from the head into the spine (9). In cases with a valve the CSF pressure did not equalise rapidly and dissociation was visible after a single cough and often more marked after a sequence (Fig. 4).

Straining: After a VALSALVA's manoeuvre produced by blowing (Figs. 2, 3) there is usually an increase in cardiac output and blood pressure with a bradycardia (Post Valsalva Rebound) and if this occurred when the CHIARI malformation was tightly impacted then the rise in CSF pressure caused by the increased pulsation in the cerebral vascular bed might be only in the head. This exacerbated the dissociation and pressure differences over 30 mm Hg were seen. This post VALSALVA rebound (PVR) was frequently associated with pounding headache for from 2 seconds to 30 seconds after straining a characteristic complaint accompanying CHIARI type I malformation.